

## Recommended clinical surveillance of patients with CNC

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### 1. Post-pubertal pediatric and adult patients:

- Echocardiogram (annually or biannually for adolescent patients with a history of excised myxoma)
- Testicular ultrasound (annually)
- Thyroid ultrasound (baseline examination; it may be repeated, as needed)
- Transabdominal ultrasound of the ovaries (baseline examination; it may be repeated, as needed)
- Urinary free cortisol levels (annually)
- Serum IGF-1 levels (annually)

### 2. Pre-pubertal pediatric patients:

- Echocardiogram (annually; *biannually* for patients with a history of excised myxoma)
- Testicular ultrasound for boys; close monitoring of growth rate and pubertal staging (annually)

### 3. Further evaluation of patients of all age groups, as needed:

For primary pigmented nodular adrenocortical disease, in addition to urinary free cortisol levels:

- Diurnal cortisol levels (11.30 pm, 12.00MN and 7.30 am, 8.00 am sampling)
- Dexamethasone-stimulation test (modified Liddle's test, as per Stratakis *et al.* [10])
- Adrenal computed tomography

For gigantism/acromegaly, in addition to serum IGF-1 levels:

- Pituitary magnetic resonance imaging
- 3-hour oral glucose tolerance test (oGTT)
- 90-minute TRH testing

For psammomatous melanotic schwannoma:

- Magnetic resonance imaging (brain, spine, chest, abdomen, retroperitoneum, pelvis)
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